SECTION 20 Hematology

HASANEIN GHALI

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1. The **LEAST** indicator for an underlying hematological problem is
   A. pallor
   B. fever
   C. petechial skin rash
   D. lymphadenopathy
   E. failure to thrive

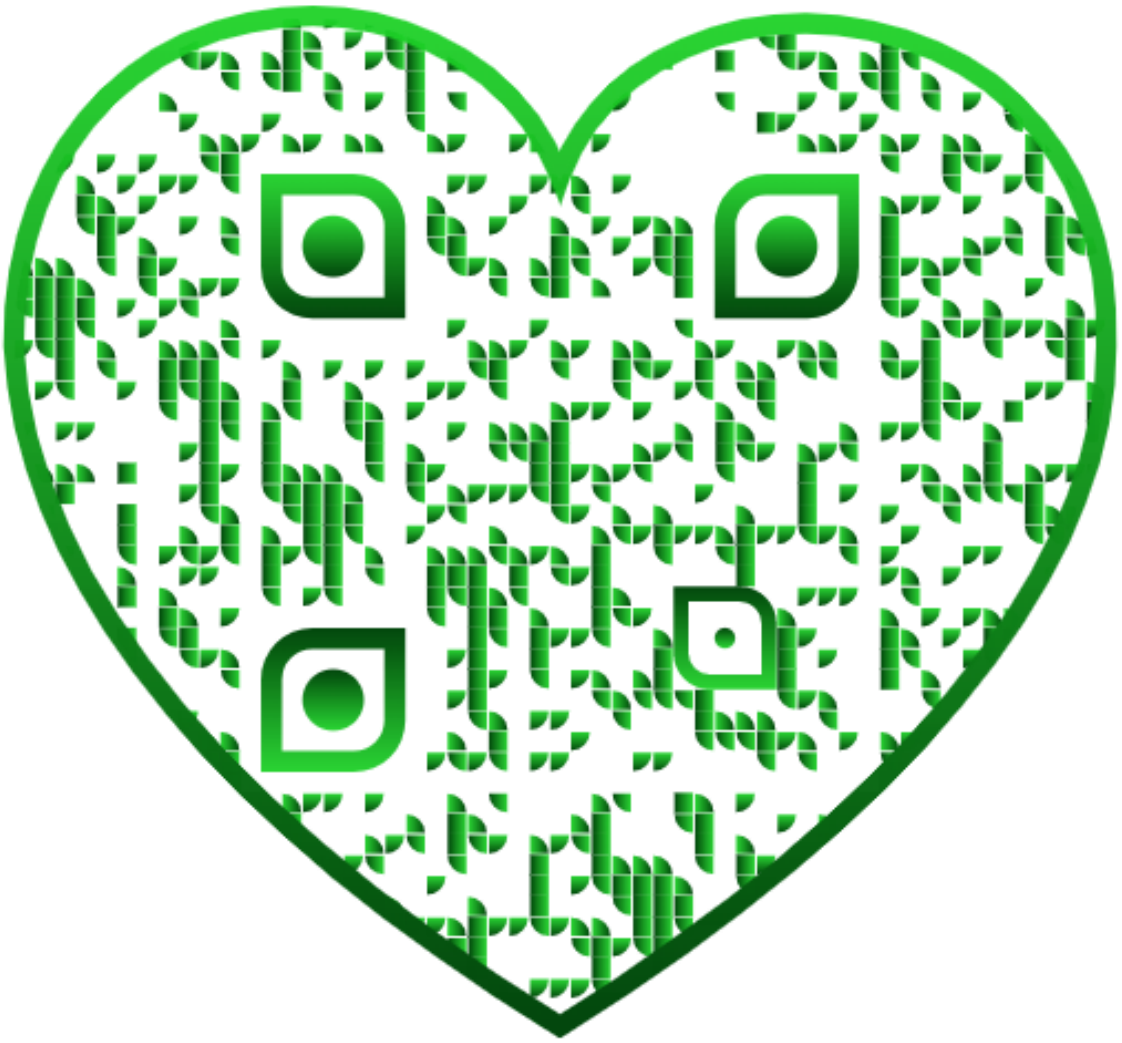
2. During intrauterine life, hematopoiesis usually begins by
   A. 3rd week of gestation
   B. 5th week of gestation
   C. 7th week of gestation
   D. 9th week of gestation
   E. 11th week of gestation

3. The primary site of hematopoiesis in the fourth week of gestational age is
   A. liver
   B. spleen
   C. lymph nodes
   D. yolk sac
   E. bone marrow

4. Which of the following symptoms/sings is a manifestation of anemia?
   A. irritability
   B. pharyngitis
   C. bruising
   D. fatigue
   E. gingivitis

5. Which of the following symptoms/signs is a manifestation of polycythemia?
   A. lymphadenopathy
   B. deep vein thrombosis
   C. epistaxis
   D. seizures
   E. oral ulceration

Irritability, cyanosis, seizures, jaundice, stroke, and headache.



6. A true hematological index for a cord blood sample is
   A. reticulocyte of 1%
   B. leukocyte of 6,000/cmm
   C. hemoglobin of 16.0 gm/dl
   D. neutrophils of 30%
   E. platelets of 90,000/cmm

Hemoglobin of 16.0 gm/dl, WBCs of 18,000/cmm, neutrophils of 60% reticulocytes of 5%.

7. Males, at onset of puberty, maintain a normal hemoglobin value about 1.5-2.0 gm/dl higher than females because of
   A. oxygen affinity of RBC is lower in males
   B. bone marrow production of RBC precursors in higher in males
   C. androgen release at the onset of puberty
   D. medullary and extramedullary site for precursors production is potent in males
   E. the RBC count is higher in males during early embryogenesis

8. An example of macrocytic anemia is
   A. thalassemia
   B. iron deficiency anemia
   C. anemia of chronic disease
   D. glucose 6-phosphate dehydrogenase deficiency
   E. folic acid deficiency

9. The urgency for packed cell transfusion in childhood anemia should be dictated by
   A. absolute level of hemoglobin
   B. extent of functional impairment
   C. age of the patient
   D. gender of the patient
   E. presence or absence of splenomegaly

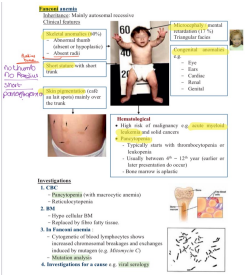
The urgency of diagnostic and therapeutic intervention, especially the use of packed RBC transfusion, should be dictated by the extent of cardiovascular or functional impairment more than the absolute level of hemoglobin.

10. The **LEAST** helpful historical clue in Iron deficiency anemia is
   A. pica
   B. age
   C. ingestion of cow's milk
   D. Giardia lamblia infection
   E. intestinal surgery

11. The **LEAST** helpful historical clue in the diagnosis of hemolytic anemia is

A. mycoplasma infection
B. gallstones
C. splenectomy of a family member
D. chronic infection
E. affected siblings

12. An 8-year-old boy presented with recurrent skin ecchymoses for the last few months. On examination; he has café-au-lait spots, microcephaly and absent thumb.



Of the following, the **MOST** likely diagnosis is
A. neurofibromatosis
B. Fanconi anemia
C. Peutz-Jeghers syndrome
D. sickle cell anemia
E. dyskeratosis congenita

13. A 5-year-old boy presented with history of frequent blood transfusion since the end of the first year of life, he has a similarly affected older sibling. On examination: he is pale and jaundiced, with frontal bossing and enlarged liver and spleen.
Of the following, the **MOST** likely diagnosis is
A. Diamond Blackfan anemia
B. thalassemia major
C. Peutz-jeghers syndrome
D. sickle cell anemia
E. dyskeratosis congenita

14. A 13-month-old girl presented with progressive pallor for the last 3 months. The mother noticed that she started to eat a non-nutritive and dirty substance. She is on breast feeding till this age. On examination, she is pale, apathetic looking, no LAP, no organomegaly, and no skin bleeds.
Of the following, the **MOST** likely diagnosis is:
A. iron deficiency anemia
B. folic acid deficiency
C. B_{12} deficiency
D. vitamin E deficiency
E. aplastic anemia

15. Vitiligo, glossitis, ataxia, and peripheral neuropathy are features of
    A. iron deficiency anemia
    B. vitamin B_{12} deficiency
    C. folic acid deficiency
    D. heavy metal intoxication
    E. thalassemia

16. Growth retardation is not evident in anemia of
    A. chronic disease
    B. malnutrition
    C. Fanconi anemia
    D. HIV infection
    E. spherocytosis → Family Hx splenomegaly cholecystectomy of gallstone

17. The individual that is **LEAST** susceptible to develop iron deficiency anemia is a
    A. 6-year- old girl with juvenile rheumatoid arthritis
    B. 13-year- old girl started menses few months ago
    C. 7-year- old girl with retinoblastoma
    D. 3-year- old boy fed large volume of cow's milk
    E. 10-month- old breast-fed infant not started solid food yet

18. The **MOST** prevalent anemia in the world is
    A. thalassemia
    B. pyruvate kinase deficiency
    C. B_{12} deficiency anemia
    D. Diamond Blackfan anemia
    E. iron deficiency anemia

19. Which percent of iron deficient children develop Iron deficiency anemia?
    A. 10%
    B. 30%
    C. 50%
    D. 70%
    E. 100%

20. Iron deficiency anemia is less prevalent in breast fed infants because
    A. breast milk contains higher amount of iron than cow's milk
    B. mothers status of iron is enough for the infant in the first year of life
    C. iron in human milk is more efficiently absorbed

D. fortification of cow's milk formula is inadequate
E. other supplements in cow's milk formula competes with iron

21. Which of the following central nervous system (CNS) abnormalities have been linked to iron deficiency anemia?
A. seizure
B. ataxia
C. poor concentration
D. blindness
E. sleep disturbance

22. The best diagnostic study for iron deficiency in an otherwise healthy child is
A. serum ferritin
B. therapeutic trial
C. serum iron
D. blood film
E. iron binding capacity

23. The earliest response to oral iron, in iron deficiency anemia is
A. improved neurologic function
good concentration يرجع
B. reticulocytosis
C. increased hemoglobin
D. reduction in iron binding capacity
E. changes in blood film

24. The increment in hemoglobin after instituting oral iron therapy for iron deficiency anemia is about
A. 1.1 gm/dl/day
B. 0.9 gm/dl/day
C. 0.7 gm/dl/day
D. 0.5 gm/dl/day
E. 0.3 gm/dl/day

25. An illogical cause of failure of increment of hemoglobin after starting treatment of iron deficiency anemia is
A. poor compliance
B. infection
C. excessive formula milk intake
D. wrong dosing

E. ongoing blood loss

26. Which of the following is **TRUE** regarding prevention of Iron deficiency anemia in infants?

A. introduction of solid foods at 3 months of age for a bottle fed infant
B. addition of iron-containing formula to breast fed infant
C. use of special high-calories formula to a bottle fed infant
D. addition of iron supplement after 6 months of age to a breast fed infant
E. introduction of solid food at 3 months of age for a breast fed infant

27. A common significant feature of blood smear in lead poisoning is

A. normochromic RBC
B. macrocytic RBC
C. basophilic stippling
D. target cells
E. raised reticulocytes

28. A feature that signifies β-halassemia minor is

A. low iron level
B. high iron binding capacity level
C. absent marrow sideroblasts
D. elevated hemoglobin A_2
E. high red cell distribution width

Hb A2 portion elevated من يكتب
thalassemia minor معناها

HbF portion elevated ومن يكتب
Hb A decrease
thalassemia major ف هاي

29. Mentzer index of less than 12.5 suggests thalassemia trait, it represents

A. MCV divided by RBC count
B. MCH divided by RBC count
C. MCHC divided by RBC count
D. PCV divided by RBC count
E. Hb divided by RBC count

30. Which of the following hemoglobinopathies is transfusion dependent?

A. alpha thalassemia trait → carrier
B. heterozygous beta thalassemia → trait
C. hemoglobin H disease → 3α Deletion
D. homozygous β-halassemia
E. sickle trait → HbA 60%. HbS 40%.

31. Which of the following results indicates Iron deficiency anemia?

Parameter	Iron deficiency anemia	α-thalassemia minor	β-thalassemia minor
MCV	↓	↓	↓
RDW	↑	Normal	Normal
RBCs	↓	Normal	Normal
Peripheral smear	Microcytosis, hypochromia	Target cells	Target cells
Serum iron studies	↓ Iron & ferritin ↑ TIBC	Normal/ ↑ iron & ferritin (RBC turnover)	Normal/ ↑ iron & ferritin (RBC turnover)
Response to iron supplementation	↑ Hemoglobin	No improvement	No improvement
Hemoglobin electrophoresis	Normal	Normal	↑ Hemoglobin A2

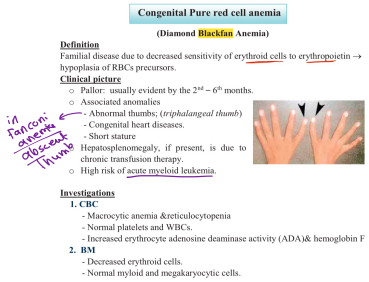
A. high serum ferritin
B. high red cell distribution width
C. increased marrow iron
D. raised marrow sideroblasts
E. reduced free erythrocyte protoporphyrin

32. Which of the following viruses can cause aplastic crises in chronic hemolytic disease?

A. Herpes virus
B. Parvovirus B19
C. Cytomegalovirus
D. Rota virus
E. T-lymphocyte virus

33. A challenging differential diagnosis of Diamond-Blackfan syndrome is

A. iron deficiency anemia
B. thalassemia syndrome
C. transient erythroblastopenia of childhood
D. spherocytosis
E. aplastic anemia


Congenital Pure red cell anemia
(Diamond Blackfan Anemia)
Definition
Familial disease due to decreased sensitivity of erythroid cells to erythropoietin → hypoplasia of RBCs precursors.
Clinical picture
Pallor: usually evident by the 2nd – 6th months.
Associated anomalies
- Abnormal thumbs; (triphalangeal thumb)
- Congenital heart diseases.
- Short stature
Hepatosplenomegaly, if present, is due to chronic transfusion therapy.
High risk of acute myeloid leukemia.
Investigations
1. CBC
- Macrocytic anemia &reticulocytopenia
- Normal platelets and WBCs.
- Increased erythrocyte adenosine deaminase activity (ADA)& hemoglobin F
2. BM
- Decreased erythroid cells.
- Normal myloid and megakaryocytic cells.

34. Which liver protein plays a key role in iron homeostasis?

A. albumin
B. fibronectin
C. opsonin
D. hepcidin
E. haptoglobin

35. The **LEAST** confirmatory finding of ABO compatibility in a 2-day- old newborn is

A. positive direct coombs test on infant's RBCs
B. positive indirect antiglobulin test on mother's serum
C. spherocytes on infant's blood smear
D. immature erythroid precursors (erythroblastosis) on infant's smear
E. elevated total serum bilirubin

36. The role of corticosteroid in autoimmune hemolytic anemia is to decrease

A. antibody production
B. the size of the spleen

C. the clearance of sensitized RBCs in the spleen
D. vascular stability
E. immune response to offending organisms

37. All the following cause autoimmune hemolytic anemia through mechanical damage of the RBCs **EXCEPT**
A. Kasabach-Merritt syndrome
B. disseminated intravascular coagulation
C. malignant hypertension
D. Mycoplasma pneumonia Cold aiha
E. hemolytic uremic syndrome

38. A 7-year-old apparently healthy boy presented to the outpatient clinic with mild pallor and slight yellowish discoloration of sclera since early childhood, family was reluctant to seek medical consultation. On examination, he had palpable spleen in addition to pallor and jaundice. His uncle had a history of repeated blood transfusions during childhood that was relieved with splenectomy.
Of the following, the **MOST** likely diagnosis at this stage is
A. autoimmune hemolytic anemia
B. hereditary spherocytosis
C. acute leukemia
D. immune thrombocytopenia
E. hemolytic uremic syndrome

39. The best treatment option for hereditary spherocytosis is
A. splenectomy
B. corticosteroids
C. immunoglobulin
D. cyclosporine
E. dapsone

40. The best clinical indicator of ongoing hemolysis in G6PD is
A. pulse rate
B. blood pressure
C. degree of pallor
D. urine color
E. respiratory rate

41. Decisions about transfusion of a child with sickle cell disease should be made on the following bases **EXCEPT**

A. age of the patient
B. clinical condition
C. hemoglobin level
D. reticulocyte count
E. presence of crises

42. The **MOST** common type of vaso-occlusive crises in children with sickle cell disease is

A. acute chest syndrome
B. pain crises

Acute chest syndrome

C. priapism
D. overt stroke
E. silent stroke

43. Parvovirus B19 infection in sickle cell disease is associated with which type of crises?

A. splenic sequestration
B. bone pain
C. priapism
D. aplastic
E. stroke

parvo – B19
pancytopnia
Aplastic anemia

44. Which of the following agents helps in decreasing the frequency of vaso-occlusive crises in children with sickle cell disease?

A. cyclosporine
B. azathioprine
C. hydroxyurea
D. tacrolimus
E. methotrexate

↑hydration
By ↑HbF

45. Beside pallor, the **MOST** common presentation of sickle cell disease during infancy is

A. dactylitis Hand-foot swelling
B. hemolytic crises
C. splenic sequestration crises
D. acute chest syndrome
E. priapism

46. Which of the following clinical conditions **DOES NOT** signify iron overload in children with β-thalassemia major?

A. heart failure
B. liver cirrhosis
C. bronzing of the skin
D. growth failure ↓IRON
E. non-immune diabetes mellitus

47. Which of the following criteria is included in the definition of severe aplastic anemia?

A. reticulocyte count less than 100.000/ µl
B. neutrophil count less than 500/mm3
C. platelets count less than 50.000/mm3
D. bone marrow cellularity less than 50% of normal
E. lymphocyte count less than 1000/mm3

48. Acquired factors that predispose to thrombosis include all the following **EXCEPT**

A. nephrotic syndrome
B. sepsis
C. abortion
D. spherocytosis
E. vasculitis

49. Which of the following conditions causes profound thrombocytopenia?

A. disseminated intravascular coagulation
B. liver failure
C. vitamin k deficiency
D. sepsis without shock
E. hemophilia A

50. In disseminated intravascular coagulation, which of the followings is reduced?

A. prothrombin time
B. partial thromboplastin time
C. D-dimer
D. fibrinogen
E. bleeding time

51. In liver failure, one of the following usually shows normal value

A. platelets count

B. prothrombin time
C. partial thromboplastin time
D. D-dimer
E. fibrinogen

52. A 6-year- old girl presented with recurrent mucocutaneous bleeding episodes since early childhood that was usually stopped spontaneously or by local measures. Her brother also complained from the same problem, initial CBC was normal.
Of the following, the **MOST** likely diagnosis is
A. hemophilia A
B. hemophilia B
C. von-Willebrand disease → ↑APTT
D. disseminated intravascular coagulation
E. immune thrombocytopenia

53. The treatment of choice for most cases of type 1 von-Willebrand disease is
A. desmopressin
B. vWF-concentrate
C. cryoprecipitate
D. platelets transfusion
E. fresh frozen plasma

54. One of the following features **DOES NOT** fit hemophilia A
A. normal ristocetin-induced platelets agglutination
B. prolonged prothrombin time APTT
C. prolonged partial thromboplastin time
D. low factor VIII activity
E. normal factor IX activity

55. One of the following features **DOES NOT** fit von-Willebrand disease
A. normal prothrombin time
B. prolonged partial thromboplastin time
C. prolonged bleeding time
D. low von-Willebrand factor activity
E. impaired platelets aggregation

56. Considering factor replacement therapy, what level of factor VIII is appropriate for mild bleeding?
A. 100%

B. 80%
C. 60%
D. 40%
E. 20%

57. One unit per kilogram body weight of factor VIII increase the plasma level by
A. 10%
B. 8%
C. 6%
D. 4%
E. 2%

58. One of the following features is **TRUE** about hemophilia inhibitors
A. IgM antibodies directed against transfused factor
B. more common in factor IX hemophiliacs
C. continuous factor infusions are the treatment of choice for high titer inhibitors
D. recombinant factor VIIa is the treatment of choice for low titer inhibitors
E. activated prothrombin complex concentrates may cause myocardial infarction

59. The **BEST** treatment option for factor VIII hemophiliacs with high titer inhibitor is
A. continuous infusion of factor VIII
B. immune modulators
C. activated prothrombin complex concentrate
D. recombinant factor VIIa
E. high dose steroids

60. A 2-year- old boy presented with recurrent skin and respiratory tract infections since early infancy, he had abnormally eczematous skin of the upper and lower limbs. His initial tests showed normal liver and renal functions with CBC showing low platelets count. He has similarly affected male cousin.
Of the following, the **MOST** likely diagnosis is
A. Glanzmann thrombasthenia
B. Wiskott-Aldrich syndrome
C. Bernard-Soulier syndrome
D. immune thrombocytopenia
E. thrombotic microangiopathy

61. A 5-year- old boy presented to the emergency department with sudden onset of skin ecchymoses and single attack of epistaxis, the history is negative for any significant event except for preceding upper respiratory tract infection. The examination showed an active healthy looking boy with diffuse skin petechial rash and ecchymoses over both lower and upper limbs, no lymphadenopathy and no organomegaly. The initial CBC is as follows: Hb 12.3 gm/dl, WBC 7.600/cmm, N 45%, L 51%, plt 17.000/cmm, and normal looking film except for rarely seen platelets. The most likely diagnosis is:

A. immune thrombocytopenia
B. acute leukemia
C. aplastic anemia
D. hemophilia A
E. von-Willebrand disease

62. All the following are treatment options for immune thrombocytopenia **EXCEPT**

A. steroids
B. androgen
C. anti-D
D. immunoglobulin
E. splenectomy

63. Splenectomy in acute immune thrombocytopenia is indicated

A. after failure of steroids therapy
B. concomitant with immunoglobulin therapy
C. in life-threatening bleeding
D. for histopathological study to exclude malignancy
E. to reduce the rate of platelet transfusions

64. Which of the following tests measures the step from fibrinogen to fibrin conversion?

A. prothrombin time
B. partial thromboplastin time
C. bleeding time
D. thrombin time
E. platelet function analyzer

65. Which of the following conditions affects partial thromboplastin time?

A. disseminated intravascular coagulation
B. thrombocytopenia

C. thrombasthenia
D. hemangioma
E. factor VII deficiency pt

66. A non-hematological cause of bleeding includes
A. hepatic failure
B. Wiskott-Aldrich syndrome
C. systemic lupus erythematosus
D. hemophilia
E. Ehlers-Danlos syndrome elasticity ↑

67. Which of the following factors affects prothrombin time?
A. Factor VII
B. Factor VIII
C. Factor IX
D. Factor XI
E. Factor XII

68. The following factors affect the common pathway **EXCEPT**
A. Factor I
B. Factor II
C. Factor V
D. Factor X
E. Factor XI intrinsic

69. Deficiency of which of the following factors is asymptomatic?
A. factor VII
B. factor VIII → significant
C. factor IX
D. factor XI mild
E. factor XII asymptomatic

70. Which of the following results correctly fits the expected transfusion outcome?
A. packed red blood cell; rise of Hb by 5-7 gm/dl
B. platelets concentrate; rise of platelets by 80.000 – 100.000/ml
C. cryoprecipitate; rise of fibrinogen by 50-100 mg/dl
D. recombinant factor VIII; rise of factor VIII by 0.5%/unit/kg
E. recombinant factor IX; rise of factor IX by 2%/unit/kg

71. One of the following factors is not available in cryoprecipitate

A. von-Willebrand factor
B. Factor XIII
C. Factor V
D. Fibrinogen
E. Factor VIII

72. A 6-year- old boy presented three hours after receiving whole blood transfusion with fever and chills, his urine color is clear and the mother stated that the transfusion process went smoothly.
Of the following, the **MOST** likely diagnosis is

A. acute hemolytic transfusion reaction
B. delayed hemolytic transfusion reaction
C. allergic transfusion reaction
D. bacteremia for the transfused blood
E. febrile non-hemolytic transfusion reaction

73. A mother called you fifteen minutes after starting blood transfusion of her 3-year- old leukemia boy for a concern of being hot, examination showed alert and vitally-stable boy with a temperature of 39.8C, his urine color is clear.
Of the following, the **BEST** treatment option now is

A. stop the transfusion
B. give antipyretics
C. check blood compatibility
D. administer hydrocortisone and diphenhydramine
E. check for any electrolyte abnormalities

74. A 6-year- old boy presented three hours after receiving blood transfusion with urticaria, pruritus, maculopapular rash, progressively developed facial edema, and respiratory distress.
Of the following, the **MOST** likely diagnosis is

A. acute hemolytic transfusion reaction
B. delayed hemolytic transfusion reaction
C. allergic transfusion reaction
D. bacteremia for the transfused blood
E. febrile non-hemolytic transfusion reaction

75. Unwise action during management of acute hemolytic reaction during blood transfusion of a 7-year- old boy is

A. stop transfusion

B. intravenous hydration
C. check compatibility
D. administer hydrocortisone and diphenhydramine
E. alkalinize urine

ياصُنّاع المُستقبل، تذكّروا إنّ
أحلامكم تَستحق منكُم عنَاء وَ كفاح..

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تمت بحمد الله وتوفيقه

SECTION 20 Hematology
ANSWERS
HASANEIN GHALI

1.**(B).** Fever is not an indicator for an underlying hematological problem. Jaundice is an indicator together with the above-mentioned distractors.
2.(**A).** Begins by the 3rd week of gestation.
3.**(D).** Hematopoiesis begins by 3 weeks of gestation with erythropoiesis in the yolk sac. By 2 months' gestation, the primary site of hematopoiesis migrates to the liver. By 5 to 6 months' gestation, the process shifts from the liver to the bone marrow.
4.**(D).** Pallor, fatigue, and heart failure.
5.**(D).** Irritability, cyanosis, seizures, jaundice, stroke, and headache.
6.**(C).** Hemoglobin of 16.0 gm/dl, WBCs of 18,000/cmm, neutrophils of 60% and reticulocytes of 5%.
7.**(C).** Androgen at onset of puberty maintain hemoglobin value about 1.5 – 2 gm/dl higher in males.
8.**(E).** Folic acid and B12 deficiency are examples of macrocytic anemias.
9.**(B).** The urgency of diagnostic and therapeutic intervention, especially the use of packed RBC transfusion, should be dictated by the extent of cardiovascular or functional impairment more than the absolute level of hemoglobin.
10.**(E).** Helpful in megaloblastic anemia.
11.**(D).** Suggests anemia of chronic disease.
12.**(B).** Fanconi anemia. Also, absent kidney, small stature, and microphthalmia.
13.**(B).**
14.**(A).**
15.**(B).**
16.**(E).** Spherocytosis usually is accompanied by normal growth.
17.**(C).** A, B, D and E are at high risk individuals.
18.**(E).** The prevalence of iron deficiency, the most common cause of anemia in the world, is about 9% in toddlers, 9% to 11% in adolescent girls, and less than 1% in teenage boys.
19.**(B).** Thirty percent of children who are iron deficient develop iron deficiency anemia.
20.**(C).** Breastfed infants are less likely to have iron deficiency than bottle-fed infants because, although there is less iron in breast milk, this iron is more efficiently absorbed.

21.**(C).** Central nervous system (CNS) abnormalities (apathy, irritability, poor concentration) have been linked to iron deficiency, presumably resulting from alterations of iron-containing enzymes (monoamine oxidase) and cytochromes.
22.**(B).** In an otherwise healthy child, a therapeutic trial of iron is the best diagnostic study for iron deficiency as long as the child is re-examined and a response is documented.
23.**(A).**The response to oral iron includes rapid subjective improvement, especially in neurologic function (within 24 to 48 hours) and reticulocytosis (48 to 72 hours); increase in hemoglobin levels (4 to 30 days); and repletion of iron stores (in 1 to 3 months).
24.**(E).** The usual therapeutic dose of 4 to 6 mg/day of elemental iron induces an increase in hemoglobin of 0.25 to 0.4 g/dL/day (a 1%/day increase in hematocrit).
25.**(C).** If the hemoglobin level fails to increase within 2 weeks after institution of iron treatment, careful re-evaluation for ongoing blood loss, development of infection, poor compliance, or other causes of microcytic anemia is required.
26.**(D).** Bottle-fed infants should receive an iron-containing formula until 12 months of age, and breastfed infants older than 6 months of age should receive an iron supplement. The introduction of iron-enriched solid foods at 6 months of age, followed by a transition to a limited amount of cow's milk and increased solid foods at 1 year, can help prevent iron deficiency anemia.
27.**(C).**
28.**(D).** This is the most important feature, others are wrong choices.
29.**(A).** If the mean corpuscular volume (MCV) divided by the RBC count is less than 12.5 (Mentzer index), the diagnosis is suggestive of thalassemia trait.
30.(**D).** Beta thalassemia major (Cooley's anemia).
31.**(B).** Low serum iron, low serum ferritin, high iron binding capacity, reduced marrow iron stores, decreased marrow sideroblasts and high free erythrocyte protoporphyrin, normal electrophoresis and high red cell distribution width.
32.**(B).** Human parvovirus B19 infects erythroid precursors and shuts down erythropoiesis.
33.**(C).** Transient erythroblastopenia of childhood, a normocytic anemia caused by suppression of RBC synthesis, usually appears after 6 months of age in an otherwise normal infant. The recovery is spontaneous.
34.**(D).** Inflammation causes an increase in the production of hepcidin, interrupting the process of iron release by macrophages and the absorption of iron from the intestines leading to anemia. Haptoglobin carries free hemoglobin released from erythrocytes.
35.**(E).** Non-specific.
36.**(C).** Corticosteroids reduce the clearance of sensitized RBCs in the spleen.

37.**(D).** Cold agglutinins.
38.**(B).** Spherocytosis is an autosomal dominant disorder but new mutations are common.
39.**(A).** Splenectomy corrects the anemia and normalizes the RBC survival in patients with hereditary spherocytosis, but the morphologic abnormalities persist.
40**.(D).** Urine color is the best clinical indicator for the ongoing hemolysis.
41.**(A).** Age of the patient is the least effective factor among the above mentioned.
42.**(B).** Pain crisis is the most common type of vasoocclusive event. The pain usually localizes to the long bones of the arms or legs but may occur in smaller bones of the hands or feet in infancy (dactylitis) or in the abdomen.
43.**(D).** In an aplastic crisis, parvovirus B19 infects RBC precursors in the bone marrow and induces transient RBC aplasia with reticulocytopenia and a rapid worsening of anemia because of the very short life span of sickle RBCs.
44.**(C).** Hydroxyurea, which increases hemoglobin F, decreases the number and severity of vasoocclusive events and frequency of acute chest syndrome in children as early as 1 year of age.
45.**(A).** Hand-foot swelling.
46.**(D).** The anemia itself is severe and leads to growth failure.
47.**(B).** Severe aplastic anemia is defined by an absolute reticulocyte count less than 50,000/μL, absolute neutrophil count less than 500/mm3, platelet count less than 20,000/mm3, and bone marrow cellularity on biopsy specimen less than 25% of normal.
48.**(D).** Indwelling catheters, vasculitis, sepsis, immobilization, nephrotic syndrome, coagulopathy, trauma, infection, surgery, inflammatory bowel disease, oral contraceptive agents, pregnancy, and abortion all predispose to thrombosis.
49.**(A).** Liver failure causes normal or reduced platelets count. The other choices do not reduce the platelet count.
50.**(D).** Fibrinogen and platelets are reduced.
51.**(A).** Sometimes reduced but usually normal.
52.**(C).** Inherited as an autosomal dominant trait and rarely as an autosomal recessive trait.
53.**(A).** Type 1 is partial deficiency of VWF and usually the deficiency is mild to moderate.
54.**(B).** The prothrombin time is normal.
55.**(E).** Platelet aggregation is normal, partial thromboplastin time is prolonged or normal and bleeding time is prolonged or normal.
56.**(D).** 40% for factor VIII is appropriate for mild episodes of bleeding whereas 80%-100% is necessary for life threatening bleeding.

57.**(E).** 1 U/kg body weight of factor VIII increases plasma level 2%, while 1.5 U/kg of factor IX increases plasma level 1%.
58.**(E).** IgG type; more common in factor VIII; high inhibitors is best treated with factor VIIa; low inhibitors is best treated with continuous factor infusion. Activated prothrombin complex concentrates, used in the past to treat inhibitor patients, paradoxically increased the risks of thrombosis, resulting in fatal complications, such as myocardial infarction.
59.**(D).** For high titer inhibitors, it is usually necessary to administer a product that bypasses the inhibitor, preferably recombinant factor VIIa.
60.**(B).** X-linked disorder characterized by hypogammaglobinemia, eczema, and thrombocytopenia caused by a molecular defect in a cytoskeletal protein common to lymphocytes and platelets.
61.**(A).** Childhood ITP is caused by an antibody (IgG or IgM) that binds to the platelet membrane.
62.**(B).** Androgen is used for Fanconi anemia.
63.**(C).** In acute ITP, only in cases of life-threatening bleeding.
64.**(D).**
65.**(A).** Hemophilia; von Willebrand disease; heparin; DIC; deficient factors XII and XI; lupus anticoagulant.
66.**(E).** Child abuse, vasculitis, telangiectasia and angiodysplasia are other causes of non-hematological bleeding.
67.**(A).** B, C, D, and E affect partial thromboplastin time.
68.**(E).** Affects the intrinsic pathway only.
69.**(E).** Factor VII, VIII and IX deficiencies cause significant bleeding, whereas factor XI deficiency causes mild bleeding.
70.**(C).** Packed red blood cell; rise of Hb by 2-3 gm/dl, platelets concentrate; rise of platelets by 30.000 – 50.000/ml, recombinant factor VIII; rise of factor VIII by 2%/unit/kg, and recombinant factor IX; rise of factor IX by 0.5%/unit/kg.
71.**(C).** Cryoprecipitate contain Factors VIII and XIII, VWF and fibrinogen.
72.**(E).** Fever during or within 4 hours after end of transfusion, chills, usually from passively transfused cytokines or recipient reaction to leukocytes. Bacteremia is a less possible cause.
73.**(B).** Treatment of febrile non-hemolytic transfusion reaction includes antipyretics for symptomatic treatment or pretreatment for future transfusions; leukocyte reduction has decreased incidence of febrile reactions. Blood culture if high suspicion for bacterial infection.
74.**(C).** Urticaria, pruritus, maculopapular rash, edema, respiratory distress and hypotension during or within 4 hours of transfusion; often because recipient has

pre-formed antibodies against donor antigens, occasionally from passive infusion of antibodies from atopic donor.

75**.(D).** Used for allergic transfusion reaction.